Cerebral palsy

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Cerebral palsy is the most common movement disorder of childhood. No decrease in prevalence of CP is reported. Improved survival of babies with disabilities may be the cause of this persistence. No specific cause can be identified for more than 50% of infants. Congenital disorders are identified in 30-40% and infections of the CNS are present in 5-10% of patients. Although there is a fixed and nonprogressive lesion, some kinds and degrees of early motor abnormalities tend to resolve without leaving significant motor disability. The treatment of CP is directed towards maximizing function and preventing secondary handicaps. The goal of therapy is to have the child to maintain his needs by himself and to adapt to environment he lives in, without being a burden to his family. An interdisciplinary team work is required to achieve this goal. [Journal of Turgut Özal Medical Center 2(1):103-106, 1995]

***Key Words** : Cerebral palsy, movement disorder*

Serebral palsi

Serebral palsi (SP), çocukluk yaş grubununun en sık görülen hareket hastalığıdır. SP'nin prevalansında düşme bildirilmemektedir. Bu devamlılıkta özürlü bebeklerin yaşam sürelerindeki iyileşme etkili olabilir. Yenidoğanların yaklaşık %50'sinden fazlasında spesifik bir neden bulunmayabilir; konjenital hastalıklar vakaların %30-40'ında, SSS infeksiyonları %5-10'unda tespit edilebilir lezyonların sabit ve nonprogressif olmalarına karşın birçok çeşit ve derecede erken motor anormallikler önemli sayılabilecek motor sakatlıklar bırakmadan çözülme eğilimindedir. SP'nin tedavi, fonksiyonları maksimum hale getirme ve sekonder handikapları önlemeye yöneliktir. Tedavinin amacı, çocuğun ailesinin yardımı olmaksızın kendi ihtiyaçlarını kendi kendine karşılama ve içinde yaşadığı ortama uyum sağlayabilmesidir. Bu amaca ulaşmak için interdisipliner ekip çalışmasına ihtiyaç vardır. [Turgut Özal Týp Merkezi Dergisi 2(1):103-106, 1995]

***Anahtar Kelimeler** : Serebral palsi, hareket hastalığı*

DEFINITION

Cerebral palsy (CP) is a term used to designate a group of conditions with a disorder of movement or posture. It results from an insult to or anomaly of the immature central nervous system (CNS).

EPIDEMIOLOGY

CP is the most common movement disorder of childhood. It occurs in 1 to 2 per 1000 children. Reports from developed countries in recent years do not suggest a decrease in the prevalence of CP.[1]

The increasing incidence noted in the past two decades is probably related to better perinatal care, especially of low birth weight (LBW) infants. This increasing incidence pattern seems to reflect improved survival of babies with disabilities.

Better treatment of Rh incompatibility states resulted in a reduction of damage to the basal ganglia. This has led to lessening of the extrapyramidal form of CP.

ETIOLOGY

Birth asphyxia is not the cause of CP in most of the children. Difficult birth in itself is merely symptom of deeper effects that influences the development of the fetus.

No specific cause can be identified for more than 50% of infants in whom the condition develops. Improved prenatal and perinatal cares have not had a favorable impact on the

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incidence of static encephalopathy which provides further support for causative factors other than birth process itself in this disorder.

Fetal bradycardia, abnormal fetal heart rate patterns, meconium in the amniotic fluid, delayed first breath and cry, low APGAR scores and low pH values are among clinical signs believed to characterize birth asphyxia. Only a very small proportion of children with these markers develop CP. For example, 99.6% of infants who had meconium in the amniotic fluid and 98% of survivors of fetal bradycardia, did not develop CP.[2]

Not all infants experiencing birth asphyxia develop CP. Only the ones showing signs of newborn encephalopathy will have CP.[3] Birth asphyxia that is significant enough to produce brain damage also injures the kidneys, liver, lungs and heart.

Abnormal antenatal events yield difficult pregnancies, labors and deliveries. Perinatal difficulties are associated with, and is not the cause of CP.

Risk factors for cerebral palsy

The conditions that augment the incidence of CP are as follows:

1. Severe birth asphyxia
2. Marked immaturity
3. LBW
4. Prematurity
5. Chorionitis
6. Marked hyperbiluribinemia
7. Toxoplasmosis and certain viral agents causing infection of CNS[4]
8. Congenital malformations
9. Multiple births
10. Placental dysfunction

No specific cause is determined in 50% of CPs. Congenital disorders are identified in 30-40% and infections of the CNS are present in 5-10% of patients. Postnatal events leading to static encephalopathy may occur throughout infancy and childhood. Of these, infection and trauma are the most significant.

PATHOLOGY

The pathologic findings that can be seen in children with CP are as follow:[5]

1. Widespread cerebral atrophy, often with cavity formation in subcortical white matter : This is a finding seen in the most severely disabled children.
2. Atrophy of basal ganglia: Children with rigidity and extrapyramidal movement disorders have this finding.
3. Atrophy and gliosis of opposite central hemisphere: This is a prominent finding encountered in hemiplegics.
4. Subependymal and intraventricular hemorrhage: This finding is observed particularly in preterms.

Gross et al. "....concluded that organic cerebral damage is often only the sequel but may even be the cause of birth complication or apparent perinatal accident."[6].

Classifications of cerebral palsy

1. **Physiologic Classification**
   - I. Pyramidal (spastic) CP : 50%
   - II. Extrapyramidal (nonspastic) CP
     - i. Choreathetoid form : 20%
     - ii. Ataxic form :
     - iii. Dystonic form : 25%
     - iv. Rigid form
   - III. Mixed CP : 5%
2. **Topographic Classification (of spastic CP)**[8]

I. Hemiplegia (25-40% of all CP cases) : Involvement of the arm and leg on one side, usually the arm affected more severely than the leg.

II. Diplegia (10-33%) : Most often associated with premature birth. Four limbs are involved. Lower extremity involvement is significant.

III. Quadriplegia (9-43%) : It is a condition with significant four limb involvement.

IV. Bilateral hemiplegia : There is spasticity of both sides of the body with significant upper extremity impairment.

3. **Functional Classification** [8,9]

Class I : There is no movement restriction.

Class II : Movement restriction is minimal or moderate.

Class III: Moderate or severe restriction of movement is present.

Class IV: No functional movement is present.

4. **Therapeutic Classification** [8,9]

Class A : There is no need for physical rehabilitation.

Class B : Minimal degree of support and habituation is needed.

Class C : Prostheses, devices, multidisciplinary team work and habituation program of long duration are needed.

Class D : Multidisciplinary team work of long

duration is required.

Members of the multidisciplinary team should consist of the following: pediatric neurologist, specialist of physical therapy and rehabilitation, audiologist, psychologist, orthopedist, physiotherapist, speech therapist, teacher of special education and the family.

DIAGNOSIS (Clinical Findings)

Diagnosis of CP usually requires documenting the presence of the following: 1. Delay in the developmental milestones, 2. Persistence of primitive reflexes, 3. Pathologic reflexes and 4. Failure to develop protective reflexes such as the parachute response.

The characteristic findings seen in pyramidal (spastic) CP are hypertonus of the clasp-knife type, hyperreflexia and pathologic reflexes. The neurologic findings of the pyramidal type are consistent and persistent, varying little with movement, tension, emotion or sleep.

The tone in extrapyramidal types varies from hypotonus to hypertonus. Extrapyramidal hypertonus is of lead-pipe type, but it is variable and changes with tension or sleep (usually hypotonic when asleep or quiet). Although pathologic reflexes are absent in extrapyramidal CP, primitive reflexes are most obvious in this type of CP.

The Summary of Clinical Findings According to the Age Groups: [9]

1. Infancy Period: Resistance to breast feeding, sucking and swallowing difficulties, excessive and meaningless crying, absence of neck control after 4 weeks of age, persistence of tonic neck reflex after 6 months, not having appropriate activity, having thumb of the hand persistently in his palm(especially after the third month) and scissoring of the legs.

2. Older Children: Delay in developmental milestones, hyperactivity, irritability, muscular hypotonicity, deficit in attention time and spasticity.

Associated neurologic abnormalities [5,7,8,10]

1. Mental retardation: Coexist in approximately 60% of patients. It is more frequent in spastic CP.
2. Communicative disorders (in the preschool child)
3. Learning disability (in the older child)
4. Deafness: Present in 10% of patients. It is associated with athetoid CP.
5. Strabismus: It is seen in 50% of patients with CP.
6. Visual defects: 25% of the hemiplegics have hemianopsia.
7. Sensory impairments: Especially present in hemiplegics.
8. Seizures: Present in 1/3rd of children with CP.
9. Behavioral disturbances (e.g. short attention span, distractibility, perseverance and self-stimulation).

PROGNOSIS

Although there is a fixed and nonprogressive lesion, some kinds and degrees of early motor abnormalities tend to resolve without leaving significant motor disability. Overall, about 50% of children who met the criteria for CP diagnosis at the age of 1 year did not have CP at school age. Resolution of clinically detected physical findings does not necessarily mean the disappearance of the underlying neuropathologic lesions. [8,11]

Monoparesis observed at 1 year of age almost always resolves; however mixed CP never resolves. Children with hemiplegia but with no other major problems almost always walk by the age of about 2 years.

More than 50% of children with spastic diplegia learn to walk by the age of 3 years. Gait is often abnormal and some children require assistive devices.

25% of children with spastic quadriplegia require total care. Approximately 33% of patients walk, usually after the age of 3 years. Most athetoid children who do not have serious accompanying spasticity walk ultimately.

Most children who sit by 2 years of age learn to walk. The children who have a Moro reflex, an asymmetric tonic neck reflex, extensor thrust and no parachute reflex are unlikely to learn to walk.

TREATMENT

The treatment of CP is directed towards maximizing function and preventing secondary handicaps. The goal of the therapy is to have the child to maintain his needs by himself and to adapt to environment he lives in, without being a burden to his family.

Physical therapy is aimed to improve motor skills, posture and locomotion, to maintain range of joint movement and to prevent contractures.

Occupational therapy is an effort to maximize self-sufficiency. Children are trained in dressing, eating and other needed skills.

Speech therapists employ auditory stimulation

and work toward improvement of muscle control for both feeding and articulation.

Drugs are used to control hypertonus, hyperactivity, involuntary movements and convulsions.

The drugs used to decrease spasticity are:

1. Diazepam (*Diazem© 2-5 mg tab*): 0.4-1 mg/kg/day, orally, in 3-4 divided doses; is used in spastic CP.
2. Baclofen (*Lioresal©, 10 mg tab*): 1-2 mg/kg/day, orally, in three divided doses; is used in spastic CP; therapeutic blood level is 82-400 ng/ml.
3. Dantrolene: 1-12 mg/kg/day, orally, in 2-4 divided doses, up to 400 mg/day; is used in spastic and athetoid children.

Also levodopa can be used in choreathetosis. The dose is 50 mg/kg/day in three divided doses.

The drugs used to decrease hyperactivity are:

1. Metifenitad (*Pitalin©*) : 0.3-1 mg/kg/day
2. Thioridazine (*Melleurettes©, 1 mg drop and 10 mg tab*) : 0.5-1 mg/kg/day in 3 divided doses.
3. Imipramin (*Tofranil©, 10-25 mg tab*) : 1.5-2 mg/kg/day in 3 divided doses.
4. Haloperidol (*Haldol©, 0.1 mg drop and 5 mg tab*) : 0.05-0.1 mg/kg/day in 2-3 divided doses.

The antiepileptic that should be selected in the children whose oral hygiene is not so well and in hyperactive children is diphenylhydantoin.

Surgical therapy can be needed to minimize the limitation of range of joint motion.

Family Counseling and Psychological Therapy

The clinician's "most important and most difficult role in providing care to children with CP is communicating with families."[12]

The problems and frustrations facing families of children with CP lead desperate families to seek nonstandard therapies. The physicians should help these families to understand the costs, hazards and uncertainties of these therapies and encourage acceptance of appropriate expectations.

If a diagnosis of CP is made, the realities of the child's future physical state can not be predicted with certainty and the requirement of the child's periodical examinations to determine the developments he will achieve, should be told to his family.

The most important of all is to avoid comments leaving the parents in an unexpectant state.

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